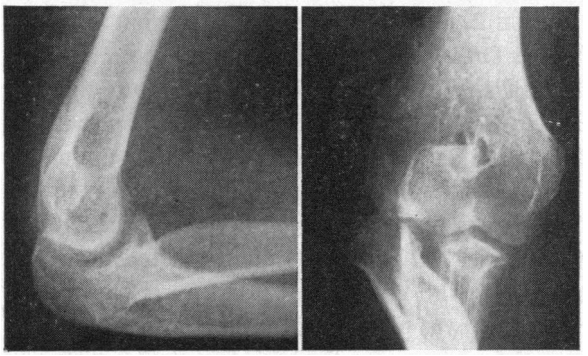

Figure 4.— (Case 2) Dislocation of radial head with greater deformity than in Case 1.

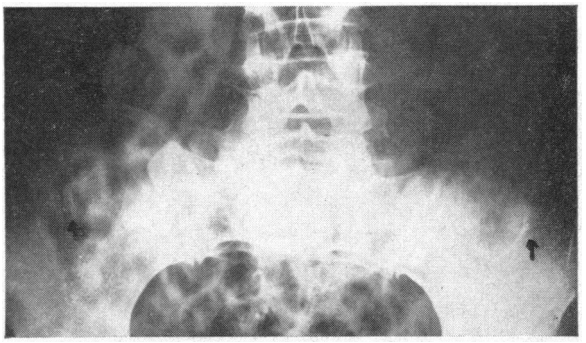

Figure 5.—Rudimentary iliac horns (arrows) in mother of patient in Case 2. (Propositus did not exhibit this manifestation.)

elbows. The mother had iliac horns as well (Figure 5).

Inquiry elicited that the maternal grandfather had involvement of the nails, knees and elbows. He had three sisters and a brother with reportedly no involvement.

Results of routine blood studies and urinalysis of the patient and her mother were within normal limits.

## Summary

The principal manifestations of hereditary osteo-onycho-dysplasia consist of nail dystrophy, elbow dysplasia, hypoplasia or complete absence of patella, and the presence of iliac horns. Nephropathic concomitants, present in some cases, may cause death.

In two cases here reported, manifestations of the condition were not present in siblings but were known in some progenitors.

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# Intrauterine Transfusion of Twins

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BEGINNING WITH the first case report of a successful fetal transfusion by Liley,[10] numerous articles have appeared in the literature describing modifications, complications and results with this procedure. The following is a report of fetal trans-

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fusions of twins in a patient with severe Rh sensitization. This case is reported because it demonstrates several problems which were encountered. We believe these problems are not unique, but experience with twin gestation, although reported in the literature, has not been outlined in detail.[5,8,9,11]

## Report of a Case

The patient was a 35-year-old, gravida 5, para 4, whose last menstrual period was 1 April 1966, and her expected date of confinement 6 January 1967. Her blood type was A, Rh negative (D), indirect Coombs (D) 1:2048. Her husband was homozygous for D. Between 1952 and 1961 the patient had four term pregnancies, all babies being born without evidence of erythroblastosis. All Rh titres during her previous pregnancies were negative. The patient's indirect Coombs titre was found, in the second trimester of this pregnancy, to be 1:1024 and she was noted to have a twin gestation. At the 32nd week of gestation she was referred to the Colorado General Hospital for evaluation. Specimens from both amniotic fluid sacs analyzed by the chloroform extraction technique[3] revealed that Twin A had an optical density (O.D.) of 0.29, and Twin B an optical density of 0.16 at 450 mu absorption peak, both in Zone III. A fetal transfusion with 75 ml of O negative packed red cells was performed without difficulty on Twin A. Before the infusion of blood, 8 to 10 ml of ascitic fluid were removed from the peritoneal cavity. Both twins did well following the first transfusion. Four days later, a specimen from the amniotic sac of Twin B revealed an O.D. of 0.46 at 450 mu, suggesting a deterioration of the condition of Twin B. Consequently, a fetal transfusion using 80 ml of packed O negative red blood cells was carried out on Twin B on 23 November 1966. Less than 5 ml of ascitic fluid was aspirated before transfusion. Although this transfusion was somewhat more difficult because the first needle introduced was put into the fetal bowel and a second transfusion needle was required to introduce the blood into the peritoneal cavity, both fetuses appeared to tolerate the procedure well.

Twenty-four hours later, fetal heart tones were heard from both fetuses. Approximately 48 hours after the second fetal transfusion, fetal movement could not be felt by the patient and fetal heart tones could not be heard in the area of Twin A but could be heard from Twin B. Ninety-six hours after the second transfusion no fetal heart tones could be heard. Ultrasound and x-ray examinations of the maternal abdomen were equivocal for fetal death. Six days after the second fetal transfusion, spontaneous labor ensued and following an uneventful labor, two macerated stillborn fetuses were delivered without difficulty. Both fetuses were females, and examination of the placenta revealed monochorionic, diamniotic membranes indicating monozygotic twins.

At postmortem examination Twin A weighed 1,970 gm. Subcutaneous tissues were decidedly edematous and the peritoneal cavity contained 50 ml of a serosanguineous fluid (adult hemoglobin). The only abnormality of the internal organs was splenomegaly. Pulmonary capillaries were engorged with nucleated red blood cells. The body of Twin B weighed 2,270 grams and there was severe generalized edema compatible with the diagnosis of hydrops fetalis. The peritoneal cavity contained 70 ml of serosanguineous fluid (adult hemoglobin) and the spleen was enlarged. Numerous nucleated red cells were noted in capillaries throughout the tissues. Examination of the 1,050-gm, edematous placenta showed findings typical of severe erythroblastosis fetalis with pale, thick, edematous cotyledons. Nucleated red blood cells were present in the vessels and there was persistence of the cytotrophoblastic cells.

## Discussion

This case presented several distinct clinical problems. In a twin gestation, accurately obtaining samples from the amniotic fluid sac of each fetus is extremely difficult. The injection of methylene blue has been recommended as a means of differentiating the two fluids.[8] However, our experience with four sets of twins has indicated that this is at best a difficult procedure. In the patient presented, ultrasound placentography indicated a left lateral implantation site. Amniocentesis in each of the four quadrants of the uterus was performed, and 450 mu O.D. absorption peak studies on each specimen of fluid showed insignificant variation ($0.29 \pm 0.04$). This suggested that only one amniotic sac had been sampled. Only after amniography was performed was a successful tap into the second amniotic cavity accomplished, revealing a 450 mu O.D. of 0.16. By outlining the extent of the amniotic cavity of Twin A, the amniogram allowed a more accurate placement of the needle through the right flank into the amniotic sac of Twin B.

In a twin gestation in which the mother shows evidence of sensitization to the D antigen, the severity of the disease may be quite different in each twin. This may be true even in monozygotic twins.[4,6] There has been no definite precedent established concerning which twin should be used as a guide to therapy. However, it has been suggested that management be governed by the most severely affected twin.[8] Because fetal transfusion carried a significant risk of fetal death, the outcome for the total pregnancy if one twin should die *in utero* had to be considered. There is evidence that the death of one twin *in utero* does not necessarily jeopardize the other.[7]

It has been established that amniotic fluid analysis is the most reliable method available for determining the severity of erythroblastosis in the fetus. From our experience with single gestations, the initial amniotic fluid values indicated that for Twin A either immediate delivery or intrauterine transfusion was required (450 mu O.D. = 0.29), while Twin B (450 mu O.D. = 0.16) could be managed conservatively by repeating the amniocentesis within seven days.[2] Because the estimated hazards of prematurity and severe erythroblastosis at 32 weeks' gestation were thought to be greater than the risks of fetal transfusion, and on the premise that management should be based on the most severely affected twin, the decision was made to transfuse Twin A. One week after the initial amniocentesis, Twin B was found have an amniotic fluid 450 mu O.D. of 0.46. Because in our laboratory this value falls in the range for fetal transfusion, with the intention to provide maximum maturity for both twins, we decided upon fetal transfusion for Twin B.

It was apparent following the amniogram of Twin A that the position of this twin changed considerably in a few hours. Consequently, we utilized the contrast material in the gastrointestinal tract to definitely identify this twin as the one requiring the first transfusion (Twin A). Purposely, an amniogram was not done on the second twin lest it cause confusion as to the identity of the untransfused twin (Twin B). We were surprised to find that the placement of the transfusion needle into the peritoneal cavity of each twin was not more difficult than has been our general experience with single fetuses of the same size.

Another but largely theoretical consideration was the possible effect of vascular anastomosis in monozygotic twins.[1] There was no way of determining either the zygosity of the twins before delivery, or, if the twins were monozygotic, the presence of vascular communications. However, it is conceivable that vascular anastomosis might be beneficial to the second twin, who thus would share in the absorption of adult red cells. On the other hand, such sharing would dilute the general effect of these cells and diminish their effectiveness for the first twin.

Finally, there was the dilemma which must be faced in many fetal transfusions in which the fetus dies *in utero*. The postmortem examination indicated successful transfusions and evidence of severe erythroblastosis fetalis with the suggestion of hydrops fetalis in each infant. However, the specific cause of death remains unknown, and whether fetal transfusion was the primary cause of death, or was a contributory factor or was completely incidental to the natural course of the disease cannot be ascertained.

## Summary

In fetal transfusion of twins in a patient with severe Rh sensitization, some of the difficulties inherent in this procedure were encountered. The value of performing an amniogram in only one twin was emphasized: (1) as an aid to accurate amniotic fluid sampling from each twin and (2) as a method to differentiate one twin from the other during the period of clinical management.

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